## 205 Serum biomarkers in CF lung disease

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This study aims to discover serum biomarkers that are sensitive enough to detect short term changes or track changes in lung disease in patients who cannot perform spirometry.

**Methods:** Serum was obtained from 22 patients at initiation and completion of intravenous antibiotic therapy for pulmonary exacerbations. A cross-sectional study included 202 patients (age 3–30 yrs) who ranged in severity of lung disease. After enrichment by affinity depletion, serum proteins were separated by 2D-gel electrophoresis (pI 5–8, MW 6–250 kD). Intensity and presence vs. absence of detected features (MCI) was compared by pair wise analyses in pre- vs. post-antibiotic samples and using regression analyses for the cross-sectional study.

**Results:** On average 1434 protein features were identified per gel. Twenty MCIs were significantly ($p < 0.005$) different in pre- versus post- therapy and were present in 90–100% of all gels. Proteins correlating with improvement in lung function were mostly inflammatory/acute phase reactants (e.g. complement C4, hemopexin). Other MCIs, which may be detected as evidence of proteolysis (e.g. Ig Lambda), decreased during therapy. Patients with clinical but insufficient pulmonary improvement showed more changes in proteins related to nutritional function (e.g. retinol binding protein). The cross-sectional study revealed 18 MCI that were present in >80% of gels and correlated with FEV1. Many of these were different MCIs than in the pre-post-study and had never been described in CF (e.g. afamin, tetranectin).

**Conclusion:** 2D-gel electrophoreses identified patterns of proteins that correlate with short term changes in lung function. Many proteins were related to inflammation but could be useful in a panel of markers to follow pulmonary improvement in patients unable to do spirometry.

## 206 Progression of lung disease on pulmonary function tests (PFT) and high-resolution computed tomography (HRCT) in children and adolescents with Cystic Fibrosis

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**Background:** In cystic fibrosis (CF), chronic bacterial infection and inflammation lead to progressive worsening in lung structure and function.

**Aim:** To compare the ability of HRCT scores and PFTs to detect changes in CF lung disease in children and adolescents with CF.

**Methods:** Baseline and follow-up HRCT scans and PFTs (at a median interval of 31 months) were retrospectively studied in a cohort of 42 CF patients (mean age: 10.9 years). Their scans were scored using 5 scoring systems (Bhalla, Hellbich, Santamaria, Brody, Oikonomou). PFTs results were expressed as percentage predicted. Chronic *Pseudomonas aeruginosa* infection and nutrition (percentage of ideal body weight – %IBW) were also assessed.

**Results:** All HRCT scores deteriorated significantly ($p < 0.0001$). FEV1 decreased by 3.5 annually ($p < 0.05$), while FVC, FEF25–75 and %IBW remained stable. Relationships between changes in HRCT scores and changes in FVC, FEV1 and FEF25–75 were weak. Baseline chronic Pseudomonas infection correlated significantly with baseline HRCT scores ($p < 0.01$), baseline FVC and FEV1 ($p < 0.05$) and with the change in FVC and FEV1 ($p < 0.05$). Substantial structural lung damage and impaired lung function was evident in some children without chronic Pseudomonas infection.

**Conclusions:** HRCT scan is more sensitive than PFTs, %IBW and chronic Pseudomonas infection in the detection of early and progressive lung disease and may be useful in the follow-up of CF patients.

## 207 Correlation of six-minute walk distance with other clinical paramaters in adult Cystic Fibrosis patients

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**Aims:** to correlate six-minute walk distance (6MWD) with other clinical parameters in adult cystic fibrosis (CF) patients.

**Methods:** six-minute walk test (6MWT) in stable adult CF patients was performed according to ATS guidelines (2002) and 6MWD was correlated with pulmonary function ($FEV_1$), chest X-ray score (Northern), clinical score (Schwachman-Kulczycki) and nutritional status (body mass index; BMI) by using Spearman correlation coefficient ($p < 0.05$ was considered as significant).

**Results:** 52 adult CF patients (30 M, 22 F) aged 25.0±4.5 years (mean±SD) were examined. 6MWD (615±61 m) was significantly correlated with $FEV_1$ (64±23% pred.), Schwachman clinical score (75±15), Northern chest X-ray score (8±3) and BMI (20.5±2.1 kg/m$^2$), as shown in Table 1.

**Consclusions:** 6MWT is useful exercise test in adult CF patients and 6MWD is significantly correlated with other clinical parameters.

**Table 1. Correlation of 6MWD with other clinical parameters**

| Parameter | $r_S$ | p |
|---|---|---|
| $FEV_1$ | 0.617 | <0.001 |
| Schwachman–Kulczycki score | 0.774 | <0.001 |
| Northern score | –0.619 | <0.001 |
| BMI | 0.368 | 0.008 |

## 208 Repeatability of cardiorespiratory measurements recorded by the LifeShirt

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The LifeShirt offers a new non-invasive method of measuring the cardiorespiratory responses to activity. Aim: To assess the repeatability of the cardiorespiratory measurements recorded by the system.

**Methods:** 20 stable patients with CF (G1) and 20 age matched healthy individuals (G2) were recruited [G1; 10M, age: 26 (9) yrs; G2; 10M, age: 24 (5) yrs]. All subjects wore the system during an endurance exercise test (6MWT) and a peak exercise test (modified shuttle test) on two occasions at least one week apart. Data relating to various cardiorespiratory parameters including: ViVol (inspiratory tidal volume); $V_e$ (minute ventilation); RR (respiratory rate); heart rate and $SpO_2$ for the 6MWT (mean values) and the modified shuttle test (peak values) were analysed. Analysis: t tests and repeated measures ANOVA assuming compound symmetry.

**Results:** On both days performance on 6MWT and modified shuttle test was significantly greater in G2 vs G1. There was no significant difference between the days in exercise performance (6MWT or the modified shuttle test) in either group. When the groups were analysed together there were no differences in cardiorespiratory responses to exercise (Table 1).

| Outcome | Modified shuttle test Mean Dif (95% CI) | 6MWT Mean Dif (95% CI) |
|---|---|---|
| ViVol (mL) | –102.6 (–433 to 228) | –23.42 (–74.6 to 27.75) |
| $V_e$ (L/min) | –4.83 (–19.32 to 9.65) | –0.12 (–2.01 to 1.78) |
| RR (breaths/min) | –1.35 (–4.27 to 1.56) | 0.08 (–0.59 to 0.76) |
| Heart rate (beats/min) | –9.37 (–19.6 to 0.86) | 1.95 (–0.40 to 4.30) |
| $SpO_2$ (%) | 3.13 (–7.34 to 13.59) | 0.21 (–1.83 to 2.26) |

**Conclusion:** This study suggests that the cardiorespiratory measurements recorded by the LifeShirt system during endurance and peak exercise tests are repeatable.